Anomalous Origins of Bilateral Vertebral Arteries in a Child with Down Syndrome and Moyamoya Disease

A Case Report

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Summary

Variations in vertebral artery origin and course are well-described in the literature. The origin of right vertebral artery from the right common carotid artery is an extremely rare variant. We describe a unique case of a child with Down syndrome with variant origins of bilateral vertebral artery, an aberrant right subclavian artery and concomitant Moyamoya disease of intracranial circulation. The presence of variations of the origin and course of craniocervical arteries might have profound implications in angiographic and surgical procedures and hence it is of great importance to be aware of such a possibility.

Introduction

The vertebral artery (VA) may originate from a number of variant positions. The presence of a vertebral origin variant must be considered in patients in whom the normal origin of the VA cannot be detected. Such variations are regarded as embryonic maldevelopments and their incidence is low. We present an extremely rare variant in which the right VA originated from the right common carotid artery (CCA) and the left VA directly from the arch. This is followed by a review of literature for embryological basis of VA variants and associated anomalies.

Case Report

A four-year-old girl, a known case of Down syndrome with mental retardation, presented with left sided hemiparesis of one year's duration. She was evaluated elsewhere initially and was being managed conservatively with some recovery of her neurodeficits. Three months into the illness, the patient also developed right upper and lower limb weakness for which she was referred to our hospital - a tertiary neurocare centre. On detailed imaging evaluation with MRI at our institute she was detected to have bihemispheric infarcts and multiple Moyamoya-like vessels. Diagnostic cerebral angiography was then performed for planning further treatment.

Cerebral Angiogram

The aortogram showed that the right CCA originated from the aortic arch as the first branch, followed by juxtaposed left CCA as the second, the left VA as the third, left subclavian artery (SCA) as the fourth while the right SCA originated as the fifth and last branch on the left side of the aortic arch beyond the origin of the left SCA (Figure 1). The right VA originated from the right CCA (Figure 2).

Selective bilateral internal carotid artery (ICA) and left VA angiograms were also carried out which revealed Moyamoya disease involving anterior and posterior circulation con-

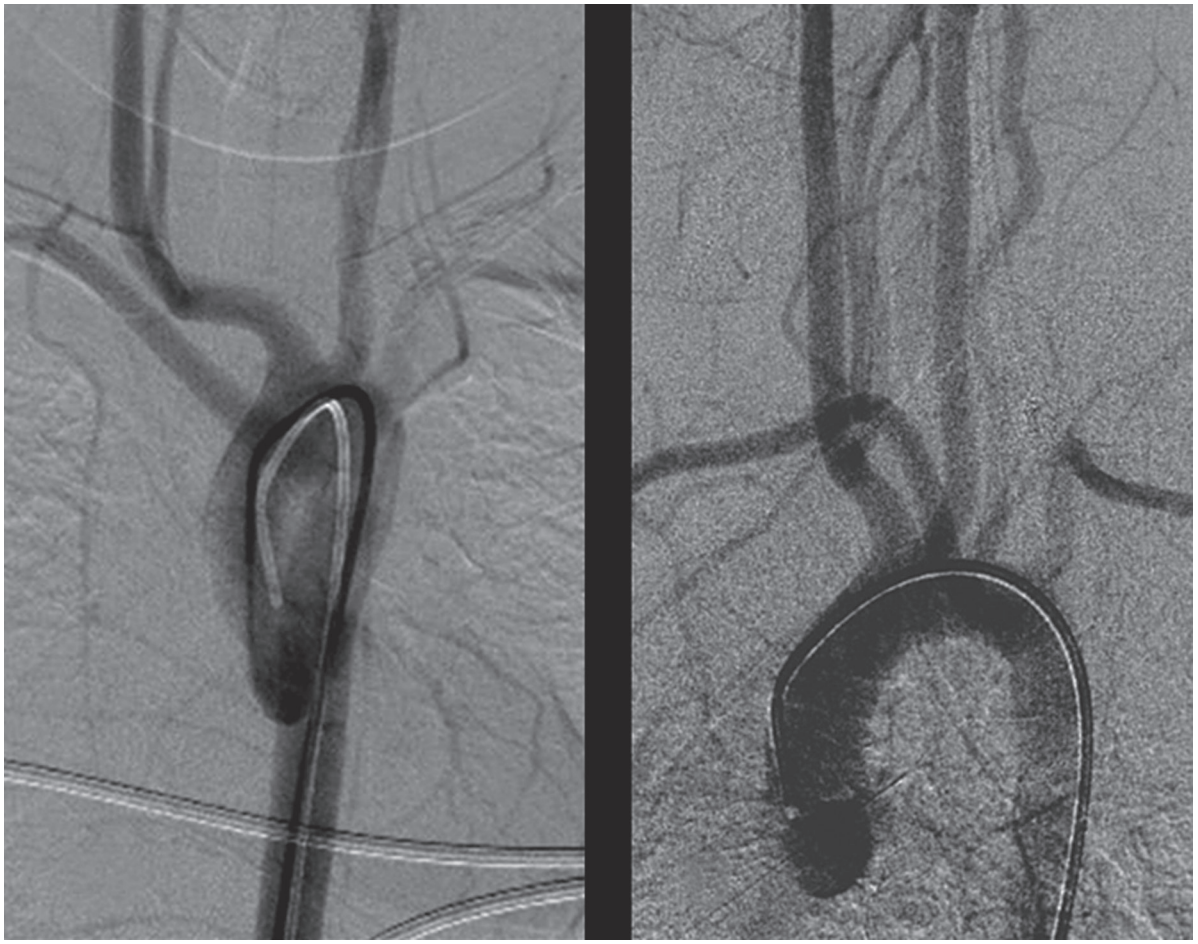

Figure 1 The common origin of right CCA & VA. A) Arch injection: The RCCA is the first branch, followed by the left common carotid artery. B) The left VA has a direct origin from the arch, followed by the left subclavian. The right subclavian artery is aberrant arising as the last branch off the arch.

sistent with Suzuki stage IV changes. Both the external carotid arteries (ECAs) were seen supplying intracranial collaterals through dural branches of external carotid artery (Figure 3).

Discussion

Although the right VA is classically described to arise as the first branch of the ipsilateral subclavian artery (SCA), multiple variations in the origin of the VA have been reported in the literature. The VA can arise from the aortic arch; from the CCA, ICA or from subclavian branches, such as the thyrocervical trunk [1-6]. Also they may have duplicate origins, generally from the aortic arch and subclavian artery and this is usually seen on the left [7]. The left VA (LVA) arises from the aortic arch with a reported prevalence of 2.4-5.8% [7]. An aberrant right SCA which arises as the last aortic branch and has a retroesophageal course, is a relatively common congenital anomaly of the aortic arch with an incidence of 0.5-1.0% [8]. On the other hand, anomalous origin of both the VA as seen in our case is not commonly reported in literature. There may be an association between such aberrant right SCA and origin of the right VA from the right CCA [1,2,6]. Tsai et al. [1] analyzed the pattern and prevalence of VA anomalies in 102 patients with aberrant SCA. They reported that 13.7% of aberrant SCA had either right VAs that originated from the right CCA or 28.6% of them had the left VA originating as the third direct branch off the aortic arch. The occurrence of anomalous origin of both VAs with aberrant right SCA, as seen in our case is very unusual.

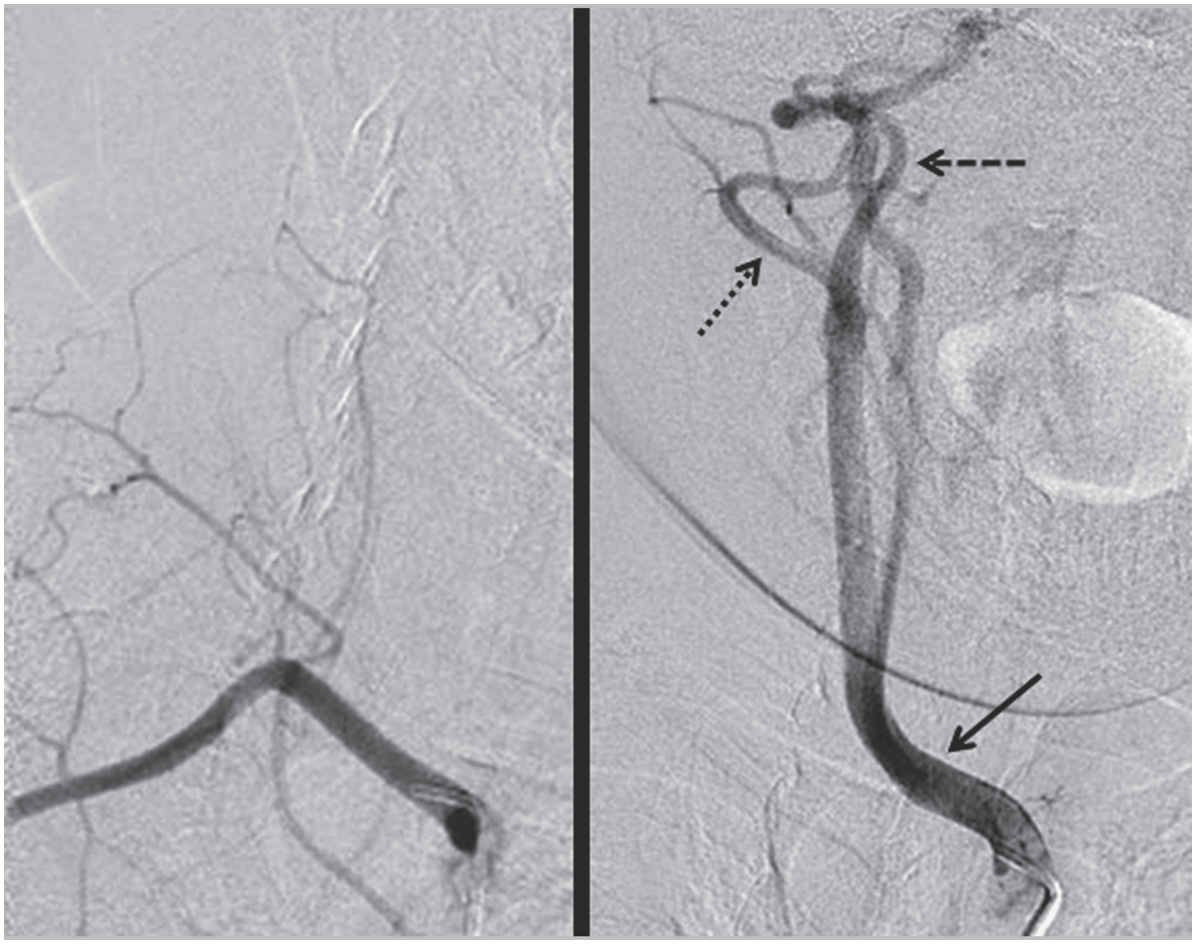

Figure 2 The RVA-CCA common origin. A) Selective right subclavian injection demonstrates the absence of VA origin. B) Selective RCCA injection demonstrates right VA (plain arrow) originating from the CCA. The CCA branches normally into ICA (dashed arrow) and ECA (dotted arrow).

Embryologically, the VA starts developing at the 7-mm stage and development is usually complete by the 14- to 17-mm stage [3]. Initially seven cervical intersegmental arteries (CIAs) originate from each of the paired dorsal aortae. Then a longitudinal anastomosis develops between the CIAs which forms the main trunk of future VAs. At the 14 to 17 mm stage, the horizontal parts of the first six CIAs disappear and the remaining seventh CIA becomes part of the subclavian artery with which the longitudinal channel communicates to form the main trunk of VA. These developmental processes result in a normal origin of the VA from the SCA. Failure of involution in one of the first six CIAs (i.e., a persistent CIA) causes a variety of abnormal origins of the VA [4,5]. If the persistent CIA occurs in the upper (first or second) intersegmental arteries, the result is an abnormal origin of the VA from the ICA or ECA; if it occurs in the lower (third through sixth) CIAs, the result is an abnormal origin of the VA from the CCA or aortic arch respectively [4,5].

Based on embryogenesis, two variants of a common origin of CCA-VA have been described [4,6]. The first (type I) includes a persistent lower CIA and an involution of the ipsilateral middle dorsal aorta between the persistent CIA and the seventh CIA. The second (type II) also has a persistent lower CIA, but instead of an involution of the ipsilateral middle dorsal aorta, the persistent CIA migrates upward to the level of the CCA. In type I anomaly, because of the persistent lower CIA, the VA comes in contact with the proximal dorsal aorta and fourth aortic arch, which in turn connects to the CCA leading to a common origin of VA/CCA. Because of the involution of the ipsilat-

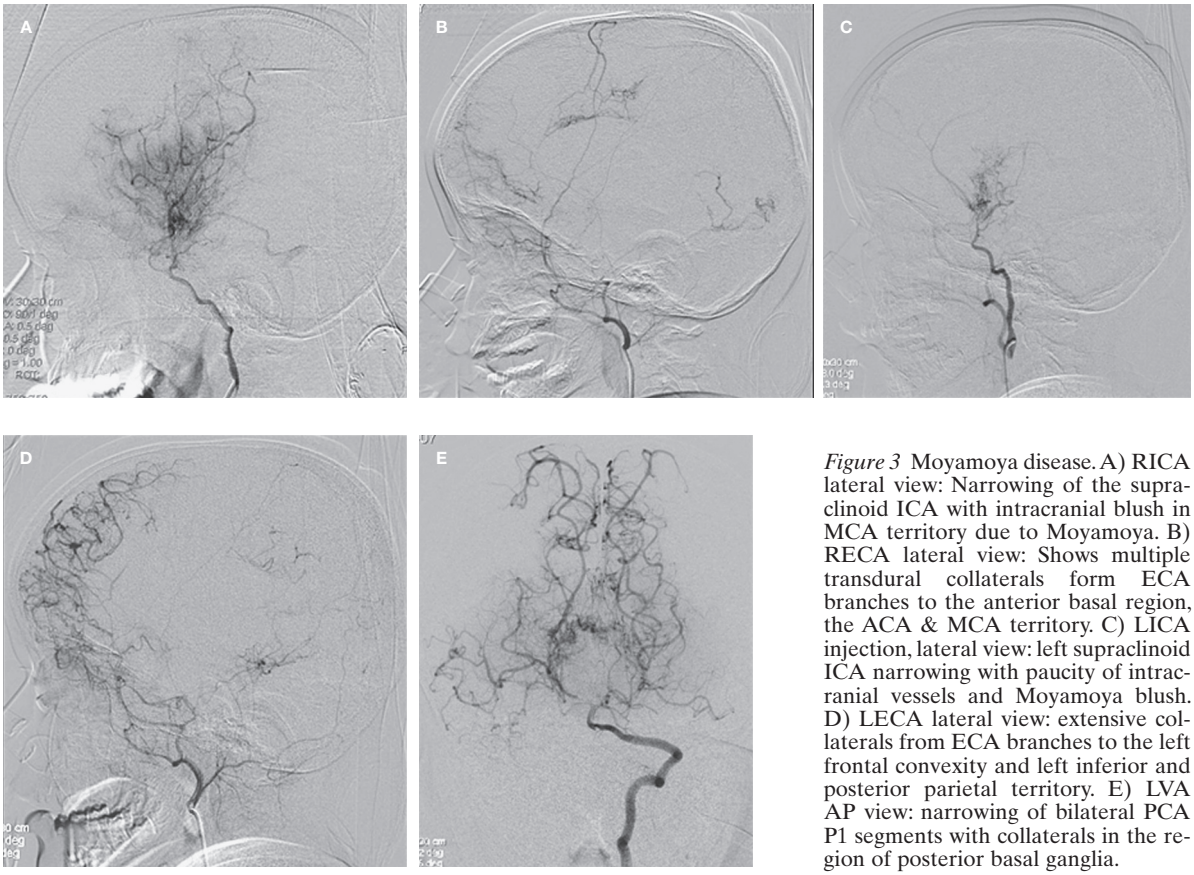

Figure 3 Moyamoya disease. A) RICA lateral view: Narrowing of the supraclinoid ICA with intracranial blush in MCA territory due to Moyamoya. B) RECA lateral view: Shows multiple transdural collaterals form ECA branches to the anterior basal region, the ACA & MCA territory. C) LICA injection, lateral view: left supraclinoid ICA narrowing with paucity of intracranial vessels and Moyamoya blush. D) LECA lateral view: extensive collaterals from ECA branches to the left frontal convexity and left inferior and posterior parietal territory. E) LVA AP view: narrowing of bilateral PCA P1 segments with collaterals in the region of posterior basal ganglia.

eral middle dorsal aorta, the remaining ipsilateral seventh CIA connects to the contralateral dorsal aorta, which results in an ipsilateral aberrant SCA [8]. Our case of right VA/CCA common origin with an ipsilateral aberrant SCA belongs to the type I anomaly.

The direct origin of LVA from aortic arch is explained by persistence of the left sixth intersegmental artery which links the longitudinal channel of LVA with aortic arch. If both the sixth and seventh intersegmental arteries persist, it leads to double origin of Left VA [5,11].

Our case had the rare combination of the following findings: anomalous origin of right VA from right CCA with associated aberrant origin of right subclavian as the last retroesophageal branch of the aortic arch, aberrant direct origin of left VA from the arch as the second branch after common CCA/VA trunk and Moyamoya pattern of intracranial circulation.

The anatomic variations of the VA are significant for diagnostic and surgical procedures in the head and neck regions. Detailed knowledge of an anomalous origin of supraaortic arteries is also of importance for patients who have to undergo four-vessel angiography in an emergency to rule out, for example, the possibility of intracranial aneurysm after subarachnoid hemorrhage. If a vertebral artery cannot be detected in the normal position, the presence of such a variant must be considered. Some authors suggested that anomalous origin of VA may lead to intracerebral disorders by altering the vascular hemodynamics, thereby placing patients at greater risk of thrombosis, aneurysm, occlusion, arterial dissection, and potentially atherosclerosis [9,10]. For cases in which the VA originates from the CCA or its branches, the ligation of the CCA may cause a compromise of the posterior fossa blood supply [10]. Anomalous VA origins also represent a potential pitfall at diagnostic cerebrovascular imaging with one or both VA being wrongly assumed to be occluded or diseased about which one needs to be careful.

In our case, the patient was also a clinically diagnosed case of Down syndrome with mental retardation. An association between Down syndrome with Moyamoya disease is well-de-

scribed [12-14] and is reported to be three times that of the general population [15]. An aberrant RSCA was detected prenatally in 16-39% of cases with Down syndrome [16]. It may be the cause of feeding difficulties in these children provided all other causes are ruled out [17]. Though the child in our case had Down syndrome, she had no feeding difficulties and the aberrant RSCA was a part of type I CCA-VA combined origin.

A case with similar arterial disposition was described by Brouwer et al. [18]. However, we report for the first time the finding of Down syndrome with Moyamoya disease and given arterial variations of arch branches as described. The incidence of Moyamoya is higher in patients with Down's syndrome compared with the general population [16], however the given arterial variations in such a patient may be incidental.

Conclusion

An interesting case of multiple anomalies of arch vessels is described. The true value of detecting anomalous origins is the diagnostic gain prior to the surgical intervention for supraaortic arteries. Awareness of variant anatomy is thus important to avoid unwarranted complications during surgical procedures and endovascular interventions. A sound knowledge of embryology helps in expecting and identifying arterial variants avoiding misinterpretation of angiographic findings.

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